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## EM-016 - ALTERED BALANCE OF PLASMA INORGANIC PYROPHOSPHATE AND SERUM TISSUE-NON-SPECIFIC ALKALINE PHOSPHATASE IN PATIENTS WITH PSEUDOXANTHOMA ELASTICUM AND CONTROLS

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### Resumen

**Objectives:** Inorganic pyrophosphate (PPi) plays a major role inhibiting dystrophic calcification. We analyze levels of PPi in patients with Pseudoxanthoma elasticum (PXE), caused by pathogenic variants in the ABCC6 gene, and controls as well as the enzymes who regulate the PPi plasma concentration.

**Methods:** We collected fasting blood samples from PXE patients and age- and sex-matched controls in EDTA and CTAD containing tubes. We measured PPi, ENPP1 mass and activity, alkaline phosphatase (AP) and tissue non-specific alkaline phosphatase (TNAP) as well CD73 by commercial enzymatic methods. Human Platelet Factor-4 (CXCL4) was assessed by ELISA.

**Results:** PPi in EDTA and CTAD samples were lower in PXE subjects than in controls ( $1.1 \pm 0.26$  vs  $1.43 \pm 0.41$  and  $0.35 \pm 0.15$  vs  $0.61 \pm 0.41$  uM/L respectively,  $p$  0.05). TNAP and liver TNAP activities were higher in PXE than in controls ( $80 \pm 14.9$  vs  $63 \pm 16$  and  $25.6 \pm 14.9$  vs  $12.9 \pm 9.2$  UI/L respectively,  $p$  0.05). ENPP1 mass and activity as well as CD73 were almost identical. There was a weak but significant inverse correlation between TNAP activity and PPi levels (Pearson correlation -0.274,  $p$  0.05) in both groups.

**Conclusions:** High TNAP activity seems to contribute to low plasma levels of PPi in subjects with PXE, reinforcing the idea that pharmacological reduction of TNAP activity may help to reduce dystrophic calcification in PXE patients.

### Bibliography

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